Nobel for UCSF scientist

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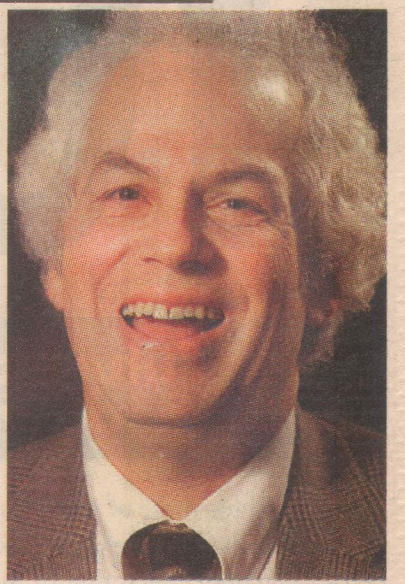
Vindication for expert widely doubted for years

By RICK WEISS
Washington Post

WASHINGTON — A maverick UC San Francisco scientist who for two decades endured derision from his peers as he tried to prove that bizarre infectious proteins could cause brain diseases like "mad cow disease" in people and animals, on Monday was awarded the ultimate in scientific vindication: the Nobel Prize in physiology or medicine.

Stanley B. Prusiner, a 55-year-old neurologist and biochemist, was cited by the Swedish Nobel committee "for his pioneering discovery of an entirely new genre of disease-causing agents and the elucidation of the underlying principles of their mode of action."

The infectious particles that Prusiner discovered, which he named prions (PREE-ons), are made of protein and do not contain any genes or genetic material — a detail that distinguishes them from all other kinds of infectious agents, such as viruses, bacteria, fungi and parasites.



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Stanley B. Prusiner, 55, said he never gave up on his research.

Until Prusiner came along, no one knew simple proteins could reproduce themselves as though they were alive. Indeed, the concept was so revolutionary that he

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was shunned for years as a man who had overreached the limits of scientific sensibility. Many researchers presumed the diseases Prusiner attributed to prions, including the sheep illness called scrapie and such human ailments as Creutzfeldt-Jakob and kuru, actually were caused by tiny, slow-growing, undiscovered viruses.

Although some scientists still question the prion hypothesis, a growing body of work from research labs around the world has led to a near-consensus that the feisty Prusiner has been correct all along.

"It's a very, very well-deserved prize," said Zach Hall, director of the National Institute of Neurological Disorders and Stroke, which funded much of Prusiner's work during the past 22 years. "The ideas were bold and the hypothesis was heretical. But he pushed unrelentingly and was unfazed."

"It's terrific," said David Baltimore, the Massachusetts Institute of Technology immunologist and 1975 Nobel laureate, who for years counted himself among Prusiner's doubters.

"These are the mythological stories of science — people who have really kept their own faith for so many years and lived through a period of opprobrium and finally are discovered to be right," Baltimore said. "Stories like these are not that common, but they provide an important lesson for young people in science: They say, "Be true to yourself.' "

In awarding Prusiner the $1 million prize, the Nobel committee of Sweden's Karolinska Institute suggested Prusiner's discoveries may lead to a better understanding of Alzheimer's disease and other neurodegenerative syndromes. His work already has proved invaluable in the study of mad cow disease — or bovine spongiform encephalopathy — and the human brain disease called "new variant" Creutzfeldt-Jakob disease, now believed to be caused by eating beef from affected cattle.

Prusiner said Monday scientists were right to doubt him at first. "I think that science should be very reticent to accept new ideas," he said. "Ninety-nine percent of new ideas are wrong. We have to be very tough on our colleagues."

But he said he gave up. "Science is about the systematic collection of data and the interpretation of that data," he said.

"You have to be forceful, you have to be aggressive, you have to be pushy. Nobody does well in science unless he has those traits. I don't know any scientists who don't push, and some push harder than others."

Prusiner learned of the award early Monday in Bethesda, Md., where he is serving on an expert committee that is advising the Food and Drug Administration about ways to reduce the spread of prion diseases.

Prions reside naturally in the brain cells of people and animals, and in their normal form they do no harm — although what exactly they do remains unclear. Occasionally, however, a prion mutates into an abnormal shape. In a peculiar process that Prusiner painstakingly documented over many years, these abnormal prions seem to manhandle surrounding normal prions into similarly defective three-dimensional forms.

As newly configured prions convert their neighbors, they multiply throughout the brain like a spreading infection. Entire pockets of brain tissue gradually die from the accumulation of abnormal proteins, leading to a "spongy" appearance characteristic of the prion brain diseases.

Prusiner also showed prion diseases sometimes can be passed from one species to another. Scientists now have strong evidence, for example, that the 21 recently discovered cases of new variant Creutzfeldt-Jakob disease in Europe were caused by the consumption of beef from cattle with mad cow disease.

Monday's award was the first time in 10 years, and only the fifth time since 1960, that the medical Nobel was awarded to just one person. Prusiner said he had no plans yet to spend the $1 million other than to pay taxes on it.